

CLINICAL VIGNETTE

Successful closure of a symptomatic left circumflex coronary artery to coronary sinus fistula

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Coronary artery fistula (CAF), first described by Krause in 1865, usually presents as an abnormal blood shunt directly from a coronary vessel into any of the cardiac chambers, great vessels, or other structures, omitting the myocardial capillaries. Although CAFs are extremely rare, with the prevalence of around 0.002% in the general population, they are one of the most common congenital coronary artery malformations and constitute up to 0.4% of all congenital cardiac anomalies. Coronary artery fistulas usually originate from the right coronary artery (50%). Left coronary origin is observed in 42% of the cases, of which left circumflex coronary artery (LCx) origin is the most common (18.3% of all CAFs observed). The majority of CAFs drain to the right ventricle (41%). Other shunts are reported to the right atrium (26%), pulmonary arteries (17%), and to the coronary sinus (CS) (7%). Although a single communication is the most common, there have been reports of multiple fistulas in a single patient.^{1,2}

Typically, CAFs are small, they are incidentally found on coronary angiography, and they do not require treatment. The clinical presentation depends on the severity of the left-to-right blood shunt. As the shunt enlarges with age, clinical symptoms may occur, such as fatigue, dyspnea, angina, endocarditis, arrhythmias, myocardial ischemia or myocardial infarction, and stroke.³⁻⁵ We report a successful surgical closure of an extremely rare, but symptomatic, LCx to CS fistula, the estimated prevalence of which is around 2 to 3 cases per 10 000 000 population.

A 61-year-old woman was urgently admitted for assessment of retrosternal pain at rest, which subsided after sublingual nitroglycerine

administration. She reported angina and fatigue on exertion for the past 10 years, hypertension, diabetes, and hyperlipidemia. Despite ST-segment depression in leads I, II, III, aVF and V₃–V₅ on admission, cardiac enzymes were negative. Transthoracic echocardiography excluded structural cardiac abnormalities, and only a mildly reduced left ventricular ejection fraction (54%) was observed. However, the treadmill stress test was positive, with ST-segment depression in leads I, II, III, and aVF with a workload of 7 METs. Computed tomography angiography was scheduled. Based on cardiac imaging, an abnormal, aneurysmatic LCx with a large fistulous connection between the LCx and CS was diagnosed (FIGURE 1A–1C, Supplementary material, Video S1). Preoperative coronary angiography confirmed the presence of the malformation and its functioning as a shunt from the left main coronary artery to the CS. Moreover, no other hemodynamically significant lesions that could explain the symptoms were observed (FIGURE 1D–1E). Therefore, due to the symptomatic presentation, coronary steal syndrome, and a large fistula diameter, which constituted a contraindication to a percutaneous intervention, the heart team referred the patient for a surgical closure of the fistula.

The surgery was performed using a median sternotomy. The malformation was intraoperatively localized deep in the myocardium, on the posterior heart wall, in the coronary sulcus. As the ventricular wall rupture in this region is usually fatal, the decision to start normothermic extracorporeal circulation and arrest the heart with crystalloid cardioplegia was made. The fistula (3 mm in diameter) was dissected deep in the myocardial tissue. It was first

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Received: August 22, 2019.

Revision accepted:

December 6, 2019.

Published online:

December 6, 2019.

Kardiologia Polska. 2019; 77 (12): 1204-1205
doi:10.33963/KP.15088

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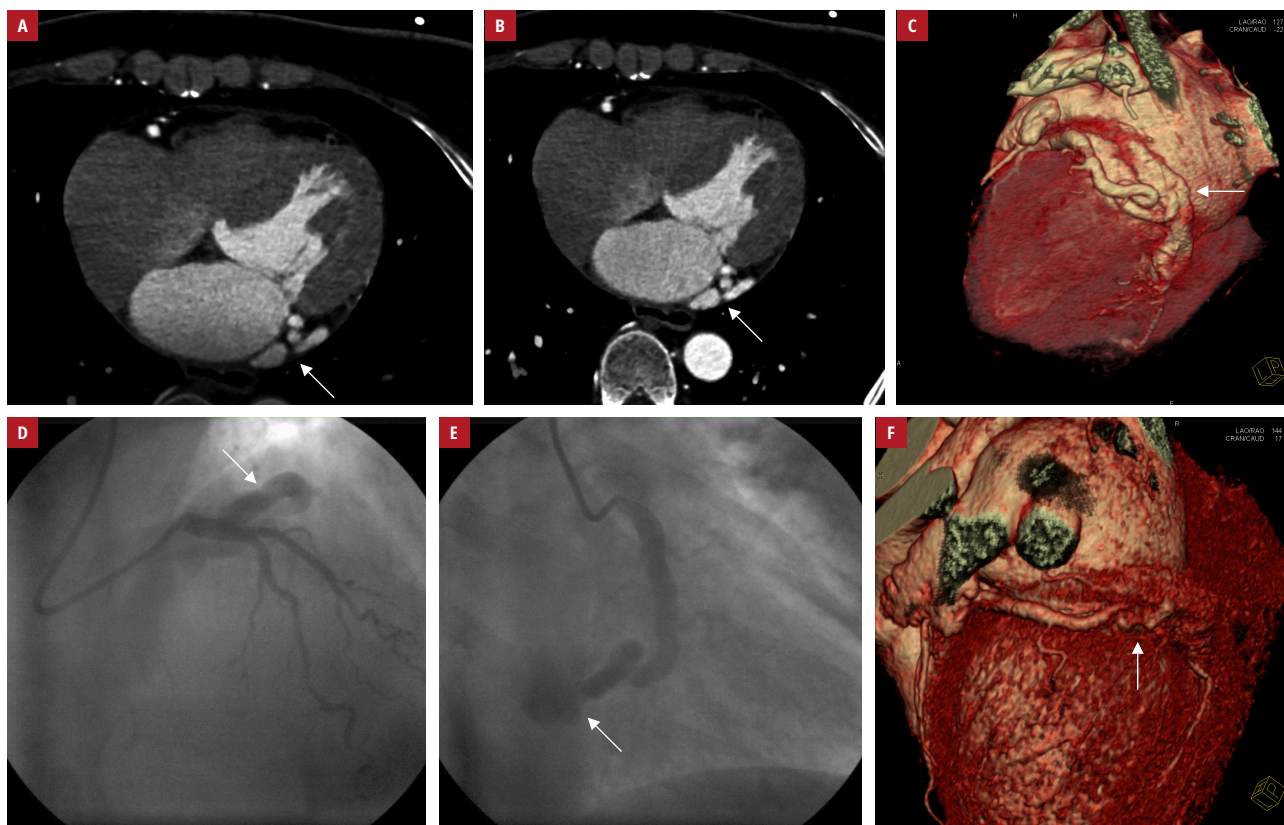


FIGURE 1 **A** – preoperative computed tomography angiography (CTA), a magnified image: left circumflex coronary artery (LCx) to coronary sinus (CS) fistula (arrow); **B** – preoperative CTA: LCx to CS fistula (arrow); **C** – preoperative CTA, 3-dimensional (3D) reconstruction: LCx to CS fistula (arrow); **D** – preoperative coronary angiogram, left main coronary artery injection: abnormal, aneurysmatic LCx (arrow); **E** – preoperative coronary angiogram, selective LCx injection with direct flow to the CS (arrow); **F** – postoperative CTA, 3D reconstruction: closure with Prolene® 5.0 pledgeted sutures (arrow)

ligated with a nonabsorbable ligature at the site of the LCx and CS connection. Then, Prolene® 5.0 pledgeted sutures (Ethicon, Bridgewater, New Jersey, United States) were added from both sides of the fistula to achieve complete occlusion and to avoid potential recanalization (FIGURE 1F). The aortic cross-clamp time was 32 minutes, and the total surgery time was 3 hours. The postoperative course was uneventful, and the patient was discharged home on postoperative day 7. The patient has not presented with any further symptoms during the 9-year outpatient follow-up.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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HOW TO CITE Samitowski Z, Mędrzycki M, Hołda MK, Kędziora A. Successful closure of a symptomatic left circumflex coronary artery to coronary sinus fistula. *Kardiologia Polska*. 2019; 77: 1204-1205. doi:10.33963/KP.15088

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